CLINICAL PROCEEDINGS

of the

CHILDREN'S HOSPITAL

WASHINGTON, D. C.

February 1951

VOLUME VII NUMBER 3

PEOPLES
DRUG STORES
INCORPORATED
AND SUBSIDIARY CORPORATIONS

WASHINGTON 2, D. C.

TO THE PROFESSION:

During January, we mailed 1951 Courtesy Cards to the members of the medical profession who requested them. These cards should have been received by now. This is the doctor's identification in our stores and if you requested a card and have not received it, please let us hear from you so we can arrange to furnish a duplicate.

We suggest that you check your supply of prescription blanks and reorder from us if needed. It is our policy to keep you supplied with these blanks, but we must depend on you to advise us of your needs.

Our prescription volume in 1950 was 2,401,530 prescriptions. This new record was only possible because of your confidence in us and we thank you for the privilege of having filled so many of the prescriptions you wrote.

Yours truly,

PEOPLES DRUG STORES, INC.

Raymond Watson

Raymond Watson
Professional Service Department

CLINICAL PROCEEDINGS

OF THE CHILDRENS HOSPITAL

13th and W Streets, Washington 9, D. C.

Vol. VII *February 1951* No. 3

CONTENTS

Published monthly by the Staff. Cases are selected from the weekly conferences held each Sunday morning at 11:00 A.M., from the Clinico-pathological conferences held every other Tuesday afternoon at 1:00 P.M., and from the monthly Staff meetings.

This bulletin is printed for the benefit of the present and former members of the Attending and Resident Staffs, and the clinical clerks of Georgetown and George Washington Universities.

Subscription rate is $1.00 per year. Those interested make checks payable to "Clinical Proceedings Dept.," The Children's Hospital, Washington, D. C. Please notify on change of address.

Entered as second class matter November 21, 1946 at the post office at Washington, D.C., under the Act of March 3, 1879. Acceptance for mailing at special rate of postage provided for in Section 538, Act of February 28, 1925, authorized January 17, 1947.

Generous IN PROTEIN

Since protein alone provides material for synthesis of new tissue, *generous quantities* of protein are needed in the infant's formula.

When LACTUM or DALACTUM is fed in the suggested amounts, the infant receives the National Research Council's Recommended Daily Allowance of protein with an additional margin for safety.

Although quantity of protein is important, *high quality* is essential, too.

All the protein of LACTUM and DALACTUM is cow's milk protein unexcelled in biologic value.

LACTUM is a whole milk and Dextri-Maltose® formula, designed for full term infants.

DALACTUM is a low fat milk and Dextri-Maltose formula, designed for prematures and full term infants with low fat tolerance.

MEADS

MEAD JOHNSON & CO.
EVANSVILLE 21, IND., U. S. A.

CANCER IN CHILDREN

Editorial

When the status quo is rather suddenly disturbed, we look about to appraise the new state of affairs which confronts us. By markedly reducing the mortality from such childhood diseases as meningitis, pneumonia, and diarrhea during the past years, such conditions as the developmental anomalies and cancer in children have assumed increasing importance. Recent advances in cardiovascular and thoracic surgery have brought a new concept and outlook for both patient and physician. Likewise, the increasing interest and knowledge, together with the improvement in the methods of diagnosis and treatment of cancer, is enabling the physician and surgeon to approach this problem with optimism instead of the feeling of hopelessness of the past.

The stimulus provided by the American Cancer Society in aiding in the organization of cancer clinics and tumor boards throughout the United States has served to make physicians and surgeons cancer conscious. During the first eighteen months of operation of the Tumor Clinic and Tumor Board at Children's Hospital, 28 per cent more patients with tumors were seen than in any other eighteen-month period in the past ten years. This fact demonstrates the value of such help.

Because of the constant reminder of its danger, the presence of cancer is being suspected and the diagnosis is made earlier than formerly. When one considers that cancer in children, leukemia included, causes more deaths than either measles, scarlet fever, epidemic meningitis, poliomyelitis, or diabetes mellitus, it will be apparent that we are confronted with a serious problem.

Malignant tumors of children are most frequently seen during the first four years of life. The same applies to leukemia which we place in the same category as cancer. The eye, kidney, bones, central nervous and lymphatic systems, and blood-forming organs are the main sites of cancer in children. Most of the malignant tumors seen in this age group are sarcomas and embryomas in contrast to carcinoma which is most frequently seen in adults. The rapidity with which metastasis takes place and the inability of many of these patients to describe their symptoms makes it necessary for the examining physician to make a thorough examination of the bones, lymph nodes, and abdomen whenever possible, if early diagnosis is to be made and effective treatment given. Three of the most common malignant tumors peculiar to children which require early surgery for cure are the retinoblastoma, the embryoma of the kidney (Wilms' tumor) and neuroblastoma.

Increasing hope for the survival or prolongation of the lives of these patients has come with better diagnostic and surgical methods, and the availability of irradiation, antibiotic and chemotherapy, and the use of the isotopes.

This group of case presentations and papers which make up this issue of *Clinical Proceedings of the Children's Hospital* attests to the increasing interest of the members of the staff in the problem of the diagnosis and treatment of cancer in infancy and childhood.

E. C. R.

ANALYSIS OF ALL TUMORS AT CHILDREN'S HOSPITAL OVER A TEN-YEAR PERIOD

Special Report No. 198

Robert H. Anderson, M.D.
M. Bruce Martin, M.D.

Few extensive pediatric tumor analyses have appeared in the literature up to the present time; as a result, the incidence of malignant tumors of infancy and childhood has not been ascertained. The frequency of malignant tumors in this age group is not realized by most physicians. As more infectious diseases are controlled by the antibiotics, the diarrheas are minimized by newer methods of treatment, and pediatric surgical problems are decimated by better surgical procedures and more competent pediatric surgery, the relative incidence, as well as the relative mortality of these tumors increases. It is imperative that these neoplasms be recognized and treated at an early stage, since they may progress rapidly to a fatal termination if not detected.

These tumors differ from those of adulthood in distribution; the usual adult malignancies of the prostate, breast, stomach, intestine, and female reproductive organs do not have to be considered. The behavior of these tumors is also in marked contrast to those of adulthood; there may be changes in the growth of the entire body or in one of its parts, changes in disposition, intelligence, or behavior. As to the cytology of these tumors, many of them probably originate from rests of embryonic cells and tend to be undifferentiated in type. Mixed tumors, embryonal tumors, and sarcomas are common while carcinomas are rare. The treatment of these tumors is also different from that of adult tumors in that growth factors must be considered, and the rapidity with which these tumors progress makes immediate treatment mandatory in many cases.

The object of this paper is to analyze all the tumors seen at Children's Hospital Washington, D. C., over a ten-year period from January 1, 1940, to January 1, 1950.

This study was undertaken primarily to compare the malignant tumors seen here during that period with tumor classifications at other institutions.

Table II compares our series with a ten-year series at Boston Children's Hospital, a series compiled by Dr. Dargeon of New York City combining seven different series, a series at the University of Chicago, and another at Chicago Children's Memorial Hospital. It is interesting to note that our

TABLE I

Total Number Admissions	71,273
Total Number Tumors	558 (0.78% of total admissions)
Total Number Malignant Tumors	144 (0.2% of total admissions)

TABLE II

INSTITUTION:	Children's Hospital Washington, D.C.	Children's Hospital Boston, Mass.	Children's Memorial Chicago, Ill.	University of Chicago, Ill.	Dr. Dargeon Combined 7 Series New York City
Total No. Cases	143	301	116	103	1770
Central Nervous System	9.8%	30.0%	13.8%	61.1%	20.8%
Lymphatic Group	49.6%	20.0%	54.3%	23.3%	11.5%
Kidney Group	17.5%	20.0%*	6.0%		13.0%
Osteogenic Group	2.1%				20.6%
Eye Group	4.2%				13.2%
Sarcoma	2.8%		6.0%	3.9%	
Neuroblastoma	7.7%		6.9%	6.8%	

* Includes neuroblastoma.

central nervous system tumors are relatively fewer than those of the other groups. Most of the brain tumors at Children's Memorial Hospital in Chicago are sent to the University of Chicago, so this may account for their large number of such tumors. Our lymphatic tumors were relatively numerous as were our kidney tumors. Some of the hospitals do not treat eye diseases, but refer them to specialized eye institutions as can also be noted. Osteogenic groups were not included in several classifications so, unfortunately, cannot all be compared but this table does give an idea of the relative incidence of these tumors. Our series can perhaps best be compared with that of Boston Children's Hospital since they were collected over a similar decade.

We have divided the tumors considered in this paper into the six main body areas where they commonly appear in children with a miscellaneous group for any other tumors:

1. Lymphatic System.
   a. Leukemia.
   b. Hodgkin's disease.
   c. Lymphoblastoma.
   d. Lymphosarcoma.
2. Kidney.
3. Brain.
4. Eye.
5. Skin.
6. Osteogenic.
7. Miscellaneous.
8. Cysts.
9. Hematomas.

1. *Lymphatic Tumors*

a. Leukemias were by far the most common tumors encountered. It is generally believed that lymphatic forms are most commonly found in children, and this was corroborated by our series. Recent treatment with anti-folic acid drugs and cortisone has prolonged the life span of these patients considerably.

b. Hodgkin's Disease is rare in children.

c. Lymphoblastoma is also unusual in the pediatric age group.

TABLE III

Leukemias—65 Cases

White	58	Types:	
Colored	7	Lymphatic	47
		Myelogenous	14
Male	34	Monocytic	1
Female	31	Aleukemic Leuk	1
		Undetermined	2

Others—6 Cases

Hodgkin's Disease	2 cases
Lymphoblastoma	1 case
Lymphosarcoma	3 cases

2. *Kidney Tumors*

Wilms' tumors (adenomyosarcoma) arise from the kidney where they increase in size rapidly and soon metastasize locally or into the chest. They are tumors of infancy and early childhood, and may be congenital. They are usually composed of a mixture of embryonal tissues. These tumors

must be detected early and removed as soon as possible. They are often treated with pre-operative irradiation to decrease the tumor size when the mass is difficult to remove surgically. Post-operative irradiation is also employed to the tumor site and often to the chest.

TABLE IV

Kidney—25 Cases

Wilms'	24	Papillary Adenocarcinoma	1
Female	11	White	18
Male	14	Colored	7

3. *Brain*

These tumors, unlike most of the others, do not metastasize outside the central nervous system unless they are metastatic lesions from elsewhere in the body. They are consequently classified according to their growth characteristics and their location and are difficult to divide into malignant and benign types. Dr. Paul Bucy of Chicago divides them into the following groups:

I. Metastatic Tumors.
II. Infratentorial Tumors.
  A. Tumors of the brain stem.
  B. Malignant tumors of the cerebellum.
    1. Medulloblastomas.
    2. Sarcomas of the Meninges.
  C. Ependymomas of Fourth Ventricle.
  D. Astrocytomas of the Cerebellum.
III. Supratentorial Tumors.
  A. Pineal Tumors.
  B. Tumors of Hypothalmus and Optic Chiasm.
  C. Craniopharyngiomas.

For practical purposes brain tumors in children can be divided into two groups: cerebellar astrocytomas (about 30 per cent) which can be completely removed in most cases with a permanent cure and the other brain tumors for which only temporary relief can be offered by surgery and other measures.

TABLE V

Brain—15 cases

Medulloblastoma	6		
Hemangioma	1		
Astrocytoma	3		
Glioblastoma Multiforme	3		
Craniopharyngioma	2		
Males	10	White	12
Females	5	Colored	3

4. *Eye*

Retinoblastomas are seen with relative frequency in early infancy, and, although very malignant, are curable. They rarely occur after three years of age. Unfortunately, there is often a delay before treatment is instituted and these tumors metastasize rapidly to bones, lymph nodes, and viscera. Early enucleation of the affected eye is effective with radiation of the other eye in case there is bilateral involvement.

TABLE VI

Eye—10 Cases

Retinoblastoma			6
Angioma, retina			1
Hemangioma, cornea			1
Papilloma, semilunar folds, lt. eye			1
Fibrolipoma, lt. eye			1
Male	8	White	7
Female	2	Colored	3
Average age			27 mos.

5. *Skin*

A. *Melanomas* may be benign but may become malignant in adolescence or later life. They should be removed if the nevus is black in color, is growing rapidly, or is in a location frequently traumatized.

B. *Neurofibromas* also may be benign and have a tendency to become malignant later (12 to 15 per cent). They are part of a constitutional disease with a marked, familial incidence. Other manifestations of neurofibromatosis are café au lait spots, subcutaneous nodules, submucous nodules, and visceral and osseus changes.

TABLE VII

Skin—117 Cases

86 Hemangiomas		1 Fibromyxoma	
6 Lipomas		6 Polyps	
4 Lymphangiomas		2 Neurofibromas	
3 Melanomas		1 Hygroma	
3 Papillomas		1 Fibromyoma	
3 Fibromas		1 Granuloma	
White	96	Male	36
Colored	21	Female	81

6. *Osteogenic*

A. *Osteochondromas* originate from bone or cartilage, are often multiple, and arise from the shaft of the bone near the tendon attachments. They may be either flat or pedunculated and may interfere with motion of the

extremities. These tumors may become malignant and form secondary chondrosarcomas, which are extremely malignant and resistant to x-ray.

B. *Osteogenic Sarcomas* are most common during puberty. They occur in the long bones near the epiphysis. There may be increased density of the medullary cavity by x-ray with some periosteal thickening. Later, after the soft tissue is invaded, the "sun-ray appearance" is noted. Both osteolytic and osteoblastic types of this tumor are resistant to irradiation and metastasize to the lungs.

C. *Ewing's Sarcomas* are very malignant and metastasize early to the lungs. They seem to respond well to irradiation initially but soon become resistant. (The irradiation may change the appearance of the tumor histologically.) On x-ray, a thickening of the cortex is seen with a widening of the medulla and a splitting of the periosteum to give an "onion-skin" appearance. Later, the x-ray may show a "sun-ray appearance" similar to that of osteogenic sarcoma. This tumor may also arise in the flat bones.

TABLE VIII

Osteogenic—8 Cases

Osteoma			2
Osteogenic sarcoma			3
Eosinophilic granuloma			2
Osteochondroma			1
Male	2	White	6
Female	6	Colored	2

Ages ranged from 22 mos. to 10 yrs.

Average—5.5 yrs.

7. *Miscellaneous*

Neuroblastomas are more frequent in the first five years of life. They originate from the sympathetic nervous system and so may occur where this tissue is present. They usually arise from the adrenals but may occur in the celiac plexus or in the sympathetic, peripheral, thoracic, or intracranial nerves. There is usually some local invasion with metastases to the skull (especially orbits), bones, liver, and lymph nodes.

These tumors are embryonal in type and rarely develop into benign ganglioneuromas or chromaffinomas. Diagnosis is made by biopsy and treatment is unsatisfactory at the present time.

In this miscellaneous group we have included tumors which did not come in the other categories. In a few cases no biopsy of the lesion was done because the procedure was considered hazardous. We have merely listed these tumors (e.g. primary tumors of the liver). In other instances the exact nature of the tumor was controversial or obscure so that the tumor was listed according to location (e.g. neurogenic tumor).

TABLE IX

Miscellaneous—38 Cases

Neuroblastoma	11	Teratoma	2	Fibromyxosarcoma	1
Polyp	3	Fibroma	2	Primary Tumor, Liver	1
Sarcoma	3	Ganglioneuroma	2	Neurogenic Tumor	1
Adenocarcinoma (thyroid)*	3	Rhabdomyosarcoma	1	Epulis, gum	1
Papilloma	2	Neurofibroma	2	Hygroma	1
		Pallatine Tumor	1	Glioma scalp	1
White	29			Male	9
Colored	9			Female	29

8. *Cysts*

There were 136 cysts with no malignant tumors included in this group.

TABLE X

Cysts—136 Cases

Sebaceous	22	Baker's	6
Thyroglossal	19	Ovarian	5
Branchial cleft	15	Lung	5
Dermoid	13	Kidney	2
Bone	6	Porencephalic	2
Dentigerous	2	Thyroid	2
Maxillar Follicular	1	Other Cysts	36

9. *Hematomas*—148 cases

Unfortunately, the records at this hospital nine or ten years ago were often incomplete so that these figures cannot be entirely relied upon. There are probably many tumors here that we have not been able to find. The cases of the early 1940's were also not adequately followed so that in many instances we have no idea of the final outcome and consequently have not been able to make any conclusions about the duration of life after treatment here.

SUMMARY

It is imperative that these tumors be detected early and, if malignant or premalignant, be thoroughly removed and treated with irradiation where indicated. Now early cancer detection and treatment is emphasized more to the laity than ever before. Research is being fostered by foundations like the Damon Runyon Cancer Fund and interest in the early detection of malignancies is promoted by Tumor Boards similar to the one now in operation at this hospital. Eventually these methods will be effective in lowering the mortality from malignancies. This new awareness is evident here when we see how rapidly our Wilms' tumors are operated on now (24–48 hours) as compared to the former 1–2 weeks prior to operation.

The systematic methods with which tumors are soon brought before the

*To be published, Theadore Winship, M.D.

Tumor Board and soon operated upon is a great credit to that group. In the final analysis, however, the entire problem must be approached via education of the laity. They must be impressed with the necessity for frequent physical examinations of well children and prompt investigation of any suspicious mass.

REFERENCES

1. Round Table Discussion on Tumors, Benign and Malignant. J. Peds. **30:** 6, 716–738 (June) 1947.
2. Mitchell-Nelson Textbook of Pediatrics, 5th Edition, W. B. Saunders and Company, Philadelphia, Pa.

THE DIAGNOSIS AND MANAGEMENT OF INTRA-ABDOMINAL MASSES IN INFANTS AND CHILDREN

Special Report No. 199

Marshall C. Sanford, M. D.

The problem of an infant or child with an unexplained intra-abdominal mass is one which frequently confronts the clinician. Initial detection often occurs during the course of a routine physical examination and symptoms may or may not be present. A consideration of the most logical diagnosis, the safest yet most informative diagnostic procedures to be employed, and the indicated therapy together with the ultimate prognosis all require a thorough understanding of the anatomy of the region as well as the pathological processes most often encountered in patients of the age under consideration.

The most likely diagnosis may vary depending on the location and characteristics of the mass and the age of the patient. Space does not permit a detailed description of all the conditions which give rise to abdominal masses, but a few of the more important general considerations regarding their management will be mentioned. Each case must be considered individually but a *plea is made for the conduction of a short, efficiently organized period of study prior to surgery which must be eventually undertaken in almost all of these conditions.*

A complete history should be taken and such important facts as G. I. or G. U. symptoms, recent infection, or history of injury should be obtained. The physical examination should be complete and attention must be paid to presence or absence of lymphadenopathy or masses elsewhere. Abdominal palpation should be thorough yet gentle and performed by as few observers as possible. The abdomen should be well relaxed and the

optimal time for examination is immediately after feeding or while the baby is sleeping. Valuable information regarding the size, shape, and consistency of the lesion can be gained by gentle bimanual palpation with slight upward pressure on the flanks. A rectal examination should not be overlooked and even in the newborn one can easily introduce a well-lubricated fifth finger into the rectum.

The safest diagnostic procedures should be employed and one should never lose sight of the patient's general condition. It is desirable to establish a diagnosis pre-operatively whenever possible but it should be realized that sometimes one must operate in order to establish this diagnosis. Don't let your patient die while obtaining an impressive array of diagnostic tests.

Air-contrast x-ray studies are safe and should be employed whenever possible—particularly for upper gastro-intestinal lesions. Barium or lipiodol may transform a partial obstruction into a complete one and the inspissation of these materials in the bowel may add greatly to the hazard of intestinal resection. The introduction of a catheter into the stomach or rectum may prove helpful in infants with suspected atresias and, if indicated, should be performed under a fluoroscope to prevent coiling within a blind pouch. Suitable contrast media for x-ray examination can be introduced through these tubes when desirable.

The majority of intra-abdominal masses are not malignant tumors but they should be regarded as such until proven otherwise. I thoroughly agree with Farber when he says:

> Every solid mass in an infant or child should be regarded as a malignant tumor until its exact nature is determined by histologic examination of the removed tumor (1).

The statement that children aren't just small adults is never better illustrated than by consideration of neoplasms. Benign tumors greatly outnumber malignant ones but the course of malignant neoplasms in childhood is an extremely rapid one with early metastasis and death. They may occur at any age although the incidence seems highest during the first year and between the ages of five and ten. Involvement of the gastro-intestinal tract, lungs, and endocrine glands is extremely rare during early life while it is quite common in adults. The type of tumor and the structure involved varies greatly with the age at which it occurs. There is no place for a defeatist attitude in the management of these patients and early recognition of the lesion and adequate therapy must be employed if the full possibilities of therapeutic success are to be accomplished.

Intra-abdominal masses may be considered by organ-systems or by quadrant location. Many of these, regardless of their position, may give some indication of their nature and origin by their size, shape, consistency,

and mobility. It also should be remembered that masses arising from or compressing the gastro-intestinal tract may cause obstruction, bleeding, or change in bowel habits. Congenital anomalies should be suspected during the first few months of life. Genito-urinary lesions often are silent. Hematuria or pyuria may be present but they seldom attract attention. Inflammatory lesions, regardless of their location, manifest evidence of infection such as fever, leukocytosis, and tenderness on palpation. Mesenteric, omental, or other masses attached by long stalks can be moved about the abdomen with ease and rarely cause symptoms unless compression exists.

A few of the more important gastro-intestinal masses may be mentioned briefly:

1. *Atresias* attract attention within the first forty-eight hours of life. The palpation of a mass rarely is the presenting complaint. Obstruction always is a prominent feature and usually is the factor responsible for the patient being brought to the physician.

2. *Duplications* are more common than is generally realized and they may occur throughout the intestinal tract. They always cause varying degrees of obstruction. Mobility of the mass depends on the segment of bowel to which it is attached. The most common sites are the small bowel and stomach.

3. *Polyps* are quite common and often associated with bleeding. When obstruction is present it usually is intermittent. The most common site for these lesions is the rectum and many are palpable on digital examination. When located in the small bowel, they often cause intussusception.

4. *Megacolon* may produce a mass which fills the entire left side of the abdomen, but a history of the patient's bowel habits is extremely helpful. Enemas and suitable x-ray studies establish the diagnosis.

5. *Diverticula* may occur throughout the gastro-intestinal tract and often are associated with bleeding, pain, or intussusception. The palpation of a mass may attract attention initially.

6. *Fecal Impactions* frequently are responsible for unexplained abdominal masses. They usually are palpable in the left side of the abdomen and, strangely enough, there may be no history of constipation or alteration in bowel habits. Cleansing enemas cause disappearance of the mass.

7. *Intussusception*, during the first few years of life, is responsible for a large number of abdominal masses, but the associated vomiting, bleeding per rectum, or crampy abdominal pain suggests the diagnosis. The ileocolic type is most common and in these the mass is felt as a sausage-shaped tumor in the right upper quadrant or epigastrium.

8. *Abscesses* or inflammatory lesions may be associated with any part of the intestinal tract but the appendiceal abscess with a mass palpable in the right lower quadrant is most common. Fever, leukocytosis, and tenderness suggest the infectious nature of the lesion.

The masses most frequently associated with the *genito-urinary system*

are: the Wilms' tumor of the kidney, a single cyst or polycystic kidney, developmental anomalies such as a horseshoe kidney, and hydronephrosis or hydroureter resulting from obstruction to the lower urinary tract. Unless infection is present these lesions may cause few symptoms except those resulting from pressure on adjacent structures or the manifestations of secondary renal damage. Intravenous pyelograms are most helpful in the recognition of these conditions.

Pelvic tumors arising from the uterus or ovary are uncommon in childhood, but their structure and position is readily determined by careful examination when they do exist.

The *liver* may be enlarged from systemic infection or may be the site of a primary inflammatory condition. Jaundice, hepatomegaly, movement of the mass with respiration, or hiccoughs may give additional evidence of the diagnosis. Hemangiomas, parasitic and non-parasitic cysts, or primary neoplasms are fairly common. Early exploration with resection whenever possible is indicated. Liver function tests are of limited aid.

Biliary masses are uncommon. The gall bladder is rarely palpable. Idiopathic dilatation of the common duct does occur. Although jaundice is present the diagnosis usually is made at operation.

Pancreatic cysts or tumors are almost nonexistent during early life. Cystic fibrosis is fairly common, but it rarely causes a palpable mass. The degree of pancreatic involvement or compression may be suggested by studies of the pancreatic secretions.

Retroperitoneal neurogenic tumors are relatively common. It has been stated that the neuroblastoma and the Wilms' tumor are the neoplasms most commonly encountered during early life. They cause few symptoms. On palpation they are firm, rubbery and fixed to the deeper structures.

Omental cysts or tumors do occur and their diagnosis often is not a difficult one since there are few lesions which allow such great mobility. The mass may be shifted about the abdomen with ease. Occasionally, tumors attached to other structures by long stalks may simulate these.

Mesenteric cysts present almost the same picture as the omental lesions except that their mobility is considerably less.

Para-umbilical masses such as a urachal cyst or persistent omphalomesenteric duct usually lie in the midline and extend down toward the pelvis. There may be a fistula between the bladder or bowel draining out through the umbilicus. *Dermoid tumors* involving the abdominal wall require little more than a knowledge of their existence to establish the diagnosis.

Splenomegaly may result from systemic infection or may be secondary to increased pressure in the portal venus circuit. Banti's syndrome causes splenomegaly, esophageal varices, and distension of abdominal wall veins. Subcapsular rupture of the spleen or leukemia may cause tremendous

splenic enlargement but in these the history of injury or blood studies may establish the diagnosis.

Lymphomas are common. They may cause generalized or localized lymphadenopathy. Sometimes the blood changes are diagnostic.

RECOMMENDED MANAGEMENT OF INTRA-ABDOMINAL MASSES

1. A detailed history determining the existence of nausea, vomiting, cramps, bleeding, change in bowel habits, previous injury, hematuria, difficulty in voiding, drainage from the umbilicus, or changes in the size of the mass.
2. Complete thorough physical examination with attention to gentle abdominal palpation by as few observers as possible.
3. Digital examination of the rectum; and when indicated, sigmoidoscopy.
4. A flat and erect x-ray of the abdomen.
5. Complete blood examination and urinalysis.
6. Make the period of study short and efficiently organized.
7. Air-contrast x-ray examination of the upper gastro-intestinal tract and when indicated, introduction of limited amounts of barium or lipiodol by mouth or tube.
8. Barium enema for low intestinal lesions or pelvic masses.
9. Intravenous pyelograms for genito-urinary or flank lesions.
10. Early surgical exploration with particular care in the gentle handling of tissues and the proper selection of an incision which will allow adequate exposure without sacrificing a strong abdominal wall.
11. Maintenance of fluid and electrolyte balance.
12. Intestinal intubation and decompression for lesions requiring resection of segments of the bowel.
13. When necessary, operate in order to establish the diagnosis.
14. If x-ray therapy is indicated at all, it should be instituted immediately post-operatively.

REFERENCE

1. FARBER, S.: Malignant Tumors in Early Life. Mitchell-Nelson Textbook of Pediatrics, 5th Edition.

PRIMARY SARCOMA OF THE OMENTUM

Case Report No. 200

Adrian Recinos, Jr., M. D.
Gerald A. McAteer, M. D.
David F. Bell, M. D.
E. Clarence Rice, M. D.

Primary sarcomas of the omentum are rare. Ninety-three have been reported in the literature. In children they are extremely rare, only four having been described under the age of sixteen years. The youngest previously recorded is that of Sala, quoted by Grieg, a child four years of age. Our patient, twenty months old, now would seem to be the youngest to suffer from this unusual malady.

CASE REPORT

C. A. U., a twenty month old white female, was admitted to Children's Hospital on January 21, 1948. Five days before admission she was unusually irritable and had very little appetite. The following day she had a temperature of 104.0 F. and a physician was called. He found only a pharyngitis and prescribed liquid sulfadiazine. There was no improvement. The temperature remained elevated and the throat red. The child developed difficult, grunting respirations and her abdomen became distended and tender. During the last two days at home the patient had several loose stools but did not vomit. On the last day, dullness was noted in the lower quadrants of the distended and tender abdomen and the child was sent to the hospital.

The past history revealed an uneventful birth and normal growth and development. The nutrition was excellent until five months prior to the child's illness at which time her appetite was described as changing from "ravenous" to "scanty". It remained poor, although her general nutritional appearance was not appreciably altered. Six weeks before admission to the hospital the patient was examined in the course of a routine check-up and no abnormalities were found.

The family history was not contributory. The patient's father, mother, and an eight month old sister were living and well.

Physical examination on admission revealed a well nourished and well developed white female appearing to be twenty months of age. She was acutely ill and breathing with moderate difficulty. The abdomen was moderately distended. The upper half was tympanitic while in the lower half there was evidence of free fluid and an ill-defined mass could be felt in the right lower quadrant. Aside from a diffusely reddened throat, there

were no other physical abnormalities. The heart and lungs appeared normal. The child's rectal temperature was 103.8 F.

The white blood count on admission was 28,400 and a catheterized urine specimen was normal except for "amorphous material" microscopically. Roentgenographic examination of the chest revealed the presence of an area of opacity in the first right interspace anteriorly having the appearance of an early interstitial bronchopneumonia. A flat plate of the abdomen gave the appearance of "intestinal obstruction with a mass of fluid in the peritoneal cavity."

A laparotomy was performed on the second hospital day. A large tumor was found in the right lower quadrant together with a large amount of fluid blood, some of it oozing from the tumor. During the operation the child went into shock but responded promptly to intravenous plasma and blood. Because of her precarious condition, no attempt was made to remove the mass. A biopsy was taken and the wound closed. Microscopic examination of the biopsy tissue disclosed an inflammatory reaction.

The post-operative course was poor. The intestinal obstruction persisted; the patient remained distended and vomited frequently. The temperature ranged from 102.0 F. to 104.0 F. Wangensteen suction, rectal tubes, and enemas were of little benefit. Whole blood, plasma, fluids, penicillin, and streptomycin were administered parenterally and the patient was placed in an oxygen tent for the greater part of her hospital stay.

Six days after the first operation a small drain was inserted into the peritoneal cavity through a half-inch incision in the right lower quadrant. The following day the abdomen was entered through a right rectus incision. A solid, grapefruit-sized, friable, and bleeding mass was removed from a stalk attachment in the right lower quadrant. The small bowel was distended. An enterostomy was performed and the tube brought out through a left flank stab wound. The tumor measured 10 x 10 x 5 centimeters and weighed 270 grams. It was composed of a smooth, glossy, gelatine-like, occasionally-hemorrhagic yellowish-white material. Microscopically, this material was found to consist of myxomatous-like tissue which was well vascularized and rather heavily infiltrated with leukocytes. The cellular elements were described as "variable in size and shape."

The patient continued to be obstructed, dehydrated, and feverish. Her nutrition and fluid and electrolyte requirements were maintained parenterally. She became gradually worse; developed dysuria, twitchings, and convulsive movements; and died quietly on the seventh hospital day.

NECROPSY FINDINGS

Post-mortem examination was limited to the abdomen. The body was that of a poorly nourished, blonde, white female child. The skin was pale,

dry, thin, and inelastic. The eyeballs were sunken. The abdomen was protuberant and tympanitic. It contained four recent, sutured incisions; one, a small wound to the left of the umbilicus, containing a small rubber catheter.

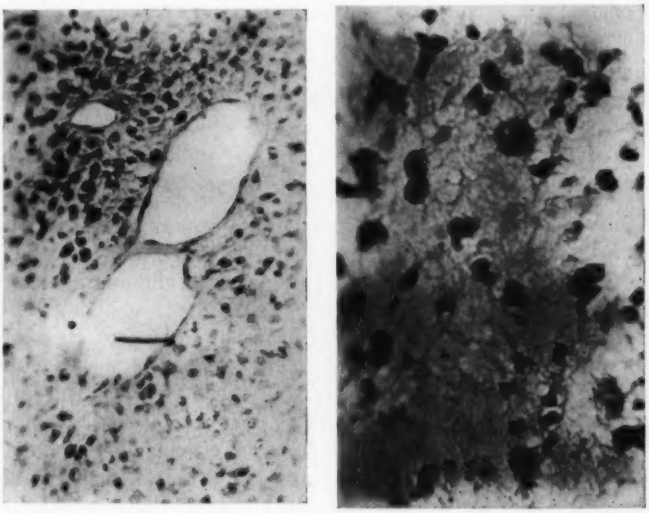
On opening the abdominal cavity, thick adhesions to the operative incisions as well as to the parietal peritoneum were noted and severed. No free fluid was present and there was no purulent exudate. All visible loops of intestine were distended and adherent to each other, to adjacent organs, and to the abdominal wall by thick adhesions. The liver had been pushed up by the dilated intestines. Indirect palpation of the lungs revealed no nodules or areas of consolidation. No mediastinal nodes could be felt and the heart did not seem enlarged.

The stomach and the first part of the duodenum were collapsed. The omentum was greatly thickened and nodular, measuring approximately 15 x 10 centimeters and varying from 0.2 to 1.5 centimeters in thickness. It was adherent to the adjacent intestinal loops and along its inferior border to the parietal peritoneum of the anterior abdominal wall. On freeing the adhesions, detaching the omentum from the stomach, and sectioning it in several places, it was noted that for a distance of 2 to 3 centimeters from its attachment to the greater curvature of the stomach the omentum was of normal appearance. Then the anterior and posterior leaves gradually fanned out enclosing a smooth, solid, occasionally-hemorrhagic tissue, most of which was gelatinous in consistency and a little of which was firm and fibrous. It was quite similar in appearance to the neoplastic tissue removed at the second operation. As the tumor was traced farther inferiorly, the enveloping leaves of the omentum could no longer be distinguished and the neoplasm extended toward the pelvis and the anterior abdominal wall in the right lower quadrant.

In the right suprapubic region there was a soft, flattened structure measuring 4 centimeters in width and dipping into the pelvis. Several catgut sutures were attached to its free border identifying it as the point from which the original tumor was removed surgically. This structure lay adjacent to the portions of the tumor along the anterior abdominal wall and may have been severed from them when the abdomen was opened. It resembled the rest of the neoplasm in appearance. This portion of the tumor was traced into the pelvis where it embraced the bladder posteriorly but spared the uterus, tubes, ovaries, and rectum. Several nodules averaging 2 to 3 centimeters in diameter were attached to the bladder posteriorly and inferiorly. It was impossible to determine grossly whether or not the bladder wall itself was invaded by these nodules.

The tumor tissue had apparently not invaded the abdominal organs and could be separated easily from the stomach and intestines to which it was attached irregularly by adhesions. There was considerable injection of most of the intestinal tract but no gross ulcerations were seen. An enterostomy

tube, partly surrounded by adhesions, was present 90 centimeters from the ileocecal valve. Distal to this tube and 34 centimeters from the cecum the ileum had doubled back on itself and the two segments were firmly bound together by adhesions. The intestinal loops proximal to this kinking were distended while the distal loops and the entire large intestine were collapsed. The mesentery and the mesenteric nodes were inconspicuous and appeared normal.



A B

FIG. 1. A and B: Fibromyxosarcoma (low and high power)

The liver was enlarged, congested, and fatty, and was not involved by the neoplastic process. The spleen was bound down by thick adhesions. There were two small accessory spleens. The kidneys, ureters, and adrenals were grossly normal.

Sections of the tumor masses were taken in and around the omentum, the bladder, internal genital organs, the peritoneal surface of the anterior abdominal wall, and the lower abdominal cavity. In spite of the variation in the components of the tumor, there was a uniformity in all the regions studied and a similarity to the tumor mass removed at operation.

Microscopic examination of numerous sections of tumor revealed it to be predominantly fibrillary in nature and well vascularized (Fig. 1). Cell borders were ill-defined or absent and the syncytium was composed of pink to grayish-pink protoplasm. The nuclei varied in size, shape and staining

qualities with occasional atypical mitotic figures. In some areas the nuclei tended to be grouped around young blood vessels. The cells varied from being moderately closely-packed in some areas to loose and edematous in others. Throughout the tissue there was a moderate infiltration of leukocytes, chiefly lymphocytes and plasma cells with occasional polymorphonuclear granulocytes. In the peripheral portions there was some necrosis with thrombosis of the vascular channels.

The tumor varied somewhat in appearance throughout the different areas examined microscopically. The inflammatory reaction was intense in some areas, altering the primary pattern of the tumor. The pattern in the more solid portions was that of a fibromyxosarcoma of low grade malignancy. The tumor had extended locally in the peritoneal cavity without evidence of distant metastases.

DISCUSSION

A review of the literature attests the rarity of primary sarcoma of the great omentum. The majority of reports have been of individual cases but occasionally they have been compiled and compared. In 1917 von Stapelmohr made a critical collection of forty-eight cases, eliminating many questionable ones. McDonald in 1927 reviewed von Stapelmohr's series and outlined the clinical and pathological features in these cases. He established strict criteria for the diagnosis of primary sarcoma of the omentum, insisting that the tumor be shown to originate in the great omentum and to be malignant by microscopic examination and clinical observation. In the past twenty years, case reports have been more frequent and several compilations of reported cases made. European authors, Grieg and Mandelstamm, and American writers: Sanes and Kenny; Ransom and Samsom; Menne and Birge; Guernsey, Kirschbaum and Teitelman; Levy and Pund; Lawler, Fox, and Cohen; and Cunningham have brought the total number to ninety-three. A trend from individual to multiple case reports is apparent in these articles suggesting that the true incidence of primary sarcoma of the omentum is greater than the reported incidence.

Throughout the years, a variety of histopathological types of primary malignant tumors of the omentum have accumulated. Levy and Pund report the relative incidence of each type as follows:

Spindle-cell sarcoma	17	Mixed-cell sarcoma	2
Fibrosarcoma	14	Endothelial sarcoma	2
Myxosarcoma	8	Perithelioma	2
Sarcoma, type not specified	7	Perivascular sarcoma	1
Round-cell sarcoma	6	Hemangio-endothelioma	1
Endothelioma	4	Myxofibrosarcoma	1
Alveolar sarcoma	3	Reticular-cell sarcoma	1
Hemangiosarcoma	3	Liposarcoma	1
Angiosarcoma	3		

This variety of terminology has brought forth differences of opinion among authorities as to the origin of malignant tumors of the omentum. Many of the tumors described are thought to have arisen from the endothelium of blood and lymph vessels. However, because these neoplasms are considered to have sarcomatous qualities, they have been grouped by the majority of the authors with the sarcomas. Guernsey, on the other hand, insists that primary malignancies of the omentum must be sarcomas because this structure is entirely mesodermal in origin. He questions the omental origin of endotheliomas, peritheliomas, endothelial sarcomas, alveolar sarcomas, etc.

Primary sarcomas of the omentum are grossly of two types: circumscribed or diffuse. The former may grow quickly to huge dimensions and may be pedunculated. The diffuse or disseminated type is more dangerous. Extension is usually by implantation metastases to near and distant parts of the peritoneum but metastases also occur in some cases (14.6 per cent in McDonald's series) outside the abdomen. These tumors have been noted frequently to extend toward the pelvis and have often been mistaken for pelvic tumors. Such was the case in our patient. Omental sarcomas are usually quite vascular and friable and contain areas of hemorrhage and necrosis with a tendency to pseudo-cysts and gelatinous degeneration. Ascites and adhesions are common and hemorrhagic ascites, considered a poor prognostic sign, occurs in one-fourth of the cases.

Although sarcoma of the omentum has been described from the age of four to eighty, the highest incidence is from twenty to fifty years and one-half of the cases are under forty. Females outnumber males by a ratio of 3 to 2.

This disease is characterized by an insidious onset of symptoms. In fact, in many cases a palpable tumor is the first evidence of illness. Abdominal pain or discomfort with a sense of increased weight in the abdomen, weakness and moderate weight loss, and abdominal distension due to ascites are the most common symptoms. The patient may also complain of anorexia, fatigue, nausea, vomiting, or constipation. Fever is not a common finding. The absence of cachexia, even in far advanced cases, has been pointed out in the literature. In our patient a marked anorexia was the only known symptom for five months. On admission to the hospital, the general nutrition of the child was noted to be good although the tumor had spread to distant parts of the abdominal cavity.

The abnormal physical findings in sarcoma of the omentum are ordinarily limited to the abdomen. A mass, often movable, can be felt in more than half of the cases and distension with evidence of fluid is nearly as common. Anemia is not an unusual finding.

Because of the rarity of this condition and the vagueness of signs and

symptoms, the clinical diagnosis is extremely difficult. Almost all reported cases were diagnosed at operation.

The prognosis is poor. An accurate survival rate cannot be deduced from the literature because the follow-up period has been extremely short in most cases. However, the mortality is high and probably exceeds 70 per cent. Many die incident to the operation and others succumb because of recurrences or metastases. The circumscribed tumors would seem to be amenable to surgical cure unless embolic phenomena have occurred from this well-vascularized structure. Pre-operative radiation therapy has been suggested but would depend on the difficult task of making an accurate diagnosis clinically.

SUMMARY

1. A case of primary sarcoma of the omentum in a twenty-month old child has been presented.

2. To the knowledge of the authors this is the youngest individual reported to be afflicted with this condition.

3. The clinical and pathological findings in this case are in agreement with those described in the literature and outlined in this report.

REFERENCES

1. Von Stapelmohr: Nord. Med. Ark., Avd. I. Kirc, 1917–18.
2. McDonald, A. L.: Sarcoma of the Omentum, Arch. Surg. **14:** 1245, 1927.
3. Grieg, D. M.: Primary Sarcoma of the Great Omentum, Edinburgh, M. J. **37:** 237 1930.
4. Mandelstamm, A.: Ueber Sarkome des grossen Hetzes, Ginek **3:** 274, 1929.
5. Ransom, H. K. and Samsom, P. C.: Malignant Tumors of the Greater Omentum. Ann. Surg, **100:** 523–534, 1934.
6. Sanes, S. and Kenny, F. E.: Primary Sarcoma of the Great Omentum, Am. J. Cancer, **21:** 795–804, 1934.
7. Menne, F. R. and Birge, R. F.: Primary Liposarcoma of the Great Omentum, Arch. Path. **22:** 823–828, 1936.
8. Levy, J. H. and Pund, E. R.: Primary Sarcoma of the Omentum: Report of 2 Cases, Am. J. Cancer, **40:** 219–226, (October) 1940.
9. Guernsey, C. M.: Primary Tumors and Cysts of the Omentum, Proc. Staff. Meet. Mayo Clin. **14:** 694 (November 1) 1939.
10. Kirschbaum, J. D. and Teitelman, S. L.: Malignant Tumors of the Greater Omentum Simulating Glomangioma, Arch. Path. **27:** 95–103.
11. Lawler, R. H., Fox, P. F., and Cohen, H.: Primary Fibrosarcoma of the Great Omentum, Am. J. Surg. **69:** 135–136 (July) 1945.
12. Cunningham, R. T.: Primary Sarcoma of the Greater Omentum, Surgery. **24:** 739–741, 1948.

WILMS' TUMOR

Case Report No. 201

Joseph M. LoPresti, M. D.
Morris Michael, M. D.
Richard L. Jackson, M. D.

The following case is reported as representing a probable cure of a Wilms' tumor, and to reemphasize to the pediatrician and the general practitioner the importance of early recognition and proper management of intra-abdominal masses in childhood.

CASE REPORT C. G. 49-184

C. G., a five week old white male, was admitted to Children's Hospital on January 4, 1949 with the chief complaint of vomiting following feedings. At the age of four weeks this infant was thought to have pyloric stenosis. At that time a small mass was palpated in the right upper abdominal quadrant. He responded dramatically to an atropine preparation. Urinary difficulty had been present almost from birth. He appeared to be uncomfortable and strained while voiding. In the two week interim before hospitalization the mass in the right upper quadrant grew rapidly in size, until on admission it measured approximately 5 centimeters in diameter.

The past history revealed that the infant weighed 5 pounds at birth. He was difficult to resuscitate, required oxygen therapy, and remained in the hospital for two weeks. Physical examination on admission was entirely negative except for the findings in the abdomen. The abdomen was distended and a large mass extending from the right costal margin to the iliac crest was palpated. This mass was not firm. Urinalysis revealed many white blood cells in the sediment, and the hemogram was normal. A flat film of the abdomen disclosed a large mass involving the entire right side of the abdomen. The intestinal contents were displaced to the left and the kidney shadows could not be demarcated. An intravenous pyelogram performed on the day of admission revealed the calyces on the right side to be markedly enlarged and displaced downward into the pelvis.

On January 5, 1949 an abdominal laparotomy was performed and a large mass weighing 145 grams was removed together with the right kidney. Microscopic examination confirmed the diagnosis of Wilms' tumor.

The post-operative course was uneventful and the infant was discharged on January 8, 1949, the fourth post-operative day. He has been followed at frequent intervals in tumor clinic, received x-ray therapy to the operative site, and has remained asymptomatic. An x-ray of the chest eighteen months after operation revealed no evidence of metastases.

DISCUSSION

Intra-abdominal masses in childhood have always presented a problem to the medical practitioner, and their management is a subject which has entailed much controversial discussion. The most common forms of intra-abdominal malignant diseases seen in the pediatric age group are the neuroblastoma and the Wilms' tumor. Both of these tumors are highly malignant and their occurrence carries a serious prognosis. They grow rapidly and metastasize early. On clinical grounds alone it is almost impossible to differentiate them, and their management, therefore, is essentially the same.

Examination of the published reports of Wilms' tumors treated in various centers indicates a high mortality rate. The greatest survival rate was reported by Silvers of the University of California[1]. In his series of eighteen patients with Wilms' tumors, thirteen had nephrectomies and ten were alive from two to fifteen years following treatment, a survival rate of 77 per cent. The largest series has been reported by Ladd and Gross[2]. With careful management and adequate follow-up they recorded a survival rate of approximately 35 per cent. Burdick et al.[3] reported twenty-four cases from the Children's Hospital, Washington, D. C. and of these patients only one could be considered cured. Since that time there have been two other cases at this hospital, one of which had a successful outcome (cf. Case Report). All authorities agree that if after one year there is no evidence of metastatic spread of the tumor, the patient may be considered cured.

The Wilms' tumor is a malignant, mixed, solid neoplasm arising in the kidney and containing many embryonal elements. It is also known as embryoma and adenomyosarcoma of the kidney. Grossly, it is rounded or slightly elongated. It is not uncommon to find areas of degeneration and hemorrhage intermixed with firmer portions. Little normal renal tissue is seen. The tendency to hemorrhage explains why these tumors enlarge rapidly and dramatically. Microscopically, the tumor is composed of epithelial masses. In some of these areas, tubular-like structures can be noted. Connective tissue is usually prominent and occasionally muscle fibers, cartilage, and bone are seen. The blood vessels may contain tumor cells. This tumor metastasizes early to the regional lymph nodes, the liver, and the lungs. This is in contradistinction to the neuroblastoma which metastasizes to the liver, long bones, and skull.

The symptomatology is conspicuous by its absence. By far the first and presenting symptom is abdominal distension or the presence of an abdominal mass. Abdominal pain, vomiting, fever, hematuria, urinary frequency, and muscle weakness may occur but are unusual. Changes in the intravenous pyelogram depend upon the location and the size of the tumor. If the tumor impinges on the kidney pelvis, distortion, fragmentation, and distension of the pelvis and calyces may occur. If the mass is of sufficient

size, the pelvis will be displaced, usually medially and downward. Similar changes can be produced by the neuroblastoma.

The Wilms' tumor must be differentiated from other abdominal masses which may occur in this age group. Among these are the neuroblastoma, hydronephrosis, polycystic kidney, intestinal duplication, and omental cyst. It should be noted that this differentiation is of academic interest only since *all of these conditions require surgical intervention.*

The management of a child who has an intra-abdominal mass is therefore predicated on the assumption that it may be of a malignant nature, namely, embryoma of the kidney or neuroblastoma. Any intra-abdominal tumor in childhood (once it has been definitely established that the mass is not an intestinal fecal impaction) is a surgical emergency. There are definite steps that should be taken in the handling of such patients:

1. Immediate hospitalization.

2. Palpation of an intra-abdominal mass should be kept at an absolute minimum. It is possible that unnecessary handling of a Wilms' tumor may force malignant cells into the blood stream and seed metastases throughout the body.

3. A basic laboratory work-up which should include a urinalysis, hemogram, flat film of the abdomen, intravenous pyelogram, and x-ray surveys of the chest, long bones, and skull.

4. After these base line studies have been carried out and anemia (if present) has been corrected, an emergency surgical laparotomy should be performed. If, at operation, the mass is found to involve the kidney, the surgeon should first ligate the renal pedicle. The retroperitoneal approach should not be used.

5. After operation, if either a Wilms' tumor or neuroblastoma has been excised, it is probably best to institute immediate and continued radiation therapy to the operative site. Radiation therapy is definitely indicated in the treatment of neuroblastoma, while it is of doubtful value in the therapy of renal embryoma because the composition of the latter may be varied, and its sensitivity to radiation is likely to vary according to the predominant cellular elements. This fact may help to explain the wide variation in results reported by different observers.

6. Frequent follow-up studies of the patient to determine the occurrence of metastases should be carried out.

The prognosis of intra-abdominal malignant disease in children is poor. In general, the younger the patient at the time of exploratory laparotomy, the better will be the results. Careful abdominal palpation in children at the time of routine physical check-ups is mandatory if these tumors are to be recognized early. It is hoped that the institution of the vigorous methods recommended will make the outlook for this small group of young patients with this type of tumor a better one.

SUMMARY

1. A Wilms' tumor with probable recovery in a five week old white male has been presented.

2. The diagnosis and management of Wilms' tumor and neuroblastoma have been discussed.

REFERENCES

1. Silvers, H. K.: J. Pediat. **31:** 643, 1947.
2. Ladd, W. E. and Gross, R. E.: Abdominal Surgery of Infancy and Childhood, W. B. Saunders Company, Philadelphia, Pa., 1947.
3. Burdick, W. F.; Jarman, W. D.; Judge, D. J.; Lattman, I.; Rice, E. C., and Sager, W.: Symposium on Wilms' Tumors, Clin. Proc. Child. Hosp. **5:** 1, 1, 1948.

CLINICO-PATHOLOGICAL CONFERENCE

Directed by: E. Clarence Rice, M.D.
Assisted by: Richard J. Waters, M.D.
Sanford Leikin, M.D.
By Invitation: Marshall C. Sanford, M.D.

Richard J. Waters, M. D.

This six year old colored female was admitted to Children's Hospital on February 23, 1946 and discharged on April 14, 1946.

The history revealed that for approximately two years this patient had been subject to frequent periods of non-productive cough lasting four to five days and occurring each month. Fever was never a factor in these illnesses; and, with the exception of associated anorexia, no other symptoms were noted. She had been treated by three practicing physicians and had made numerous visits to the out-patient clinic of this hospital. Treatment during this period was confined to expectorants and chemotherapy to which she did not respond favorably. The distressing cough continued unabated.

Three months prior to admission her parents noted a gradually progressive malaise. The patient complained of being "short of breath" and soon took little interest in the strenuous outdoor activities indulged in by her neighborhood playmates. At about this time, the coughing was accompanied by the expectoration of strings of dark red mucus. On one occasion about one-half teaspoon of bright red blood followed strenuous coughing. On the day of admission, the patient complained of mild pain in the left chest and was taken to the out-patient dispensary. She was admitted to the ward for observation and the study of suspected pulmonary pathology.

Past history revealed that she had experienced the usual childhood diseases without complication. Before the onset of the present illness two years previously, she had seldom seen a physician and was considered normal by her parents.

Family history was non-contributory and negative for tuberculosis, diabetes, and cancer.

Physical examination on admission revealed a well developed but thin colored female in no distress. Her temperature was 99.4 F. rectally; her pulse, 88; and respirations, 24. Examination of the eyes, ears, and nose was negative. The throat was moderately injected. The neck was described as normal and a few small lymph nodes were felt in the post-cervical areas. Examination of the lungs revealed normal breath sounds and percussion note in all areas. The heart was not enlarged. The rate and rhythm were normal and no murmurs were heard. No organs or masses were palpable within the abdomen. The neurological examination was negative.

A urinalysis was normal. A hemogram revealed 11.0 grams of hemoglobin; 9,100 white blood cells with 63 polymorphonuclears, 31 lymphocytes, 4 monocytes, and 1 eosinophile per 100 cells. A serologic test for syphilis was negative. Roentgenograms revealed "the presence of a high, left paramediastinal opacity." (Figs. 1 and 2.)

The patient was prepared for surgery and on March 29, 1936 an exploratory thoracotomy was performed.

DISCUSSION

Marshall C. Sanford, M. D.: In considering this case we see that we are dealing with a six-year old colored female who had a radio-opaque shadow in her chest. I think the age is an important feature since the incidence of many lesions varies considerably with the age of the patient. She was hospitalized for more than one month prior to operation and during this time we assume she was undergoing study or treatment. She was then subjected to operation and was discharged from the hospital sixteen days later. An understanding of surgical procedures will give us some indication of the type of operation that must have been performed.

This child was perfectly well for the first four years of her life but during the next two years, she had a non-productive cough which did not respond favorably to chemotherapy or expectorants. On one occasion she coughed up some bright red blood. Dyspnea was the factor responsible for her hospitalization. This began only a few weeks prior to admission. I am quite interested in the statement that she expectorated "strings of dark red mucous." If this has some hidden meaning, and the "strings" could be identified as hair, a very obvious diagnosis suggests itself.

Turning now to the physical examination, we find that it was entirely

normal. The chest was clear to percussion and auscultation. This is an important consideration and helps rule out a number of possible diagnoses. As far as the x-rays are concerned, on the AP view, there is a smooth, rounded, radio-opaque mass lying just to the left of the midline extending from the second to the fifth interspaces posteriorly. The technique of the lateral film, unfortunately, is quite poor and the mass cannot be visualized in this view. On the right anterior oblique, there appears to be a smooth, lemon-sized mass located well posteriorly in or about the mediastinum.

Was a Horner's syndrome present?

Dr. Waters: No, there was no mention of this in the record.

Dr. Sanford: In a consideration of the possibilities that might conceivably explain this shadow, the first question we must answer is: "Is this a pulmonary lesion or an extra-pulmonary one?"

I. *Pulmonary*

A. *Pulmonary cysts* may involve the parenchyma of the lung. They may be air-containing or fluid-containing. Those containing air obviously communicate with a bronchus, although this communication may be difficult to demonstrate. Those containing fluid also may have a communication with a bronchus and when they do the expectoration of this fluid occurs at times. These cysts are prone to become infected. This child was free of infection and since the shadow obviously is not an air-containing one, I feel that a pulmonary cyst is unlikely. Bronchogenic cysts do occur but the most common site is the bifurcation of the trachea. The lung findings are definitely against this.

B. *Abscesses* do occur but are more common in the lower lobes and the x-ray always reveals an area of inflammatory tissue surrounding the lesion. This appears as a halo about the center of the mass. There is no evidence of cavitation or pleural thickening. These findings together with the absence of fever or leukocytosis would tend to exclude this as a possibility.

C. *Unresolved pneumonia* is another possibility. Except for the AP view, the x-rays tend to rule this out. The course is too long and the consolidation too localized for the usual lesion of this type. The most suggestive evidence against this is the fact that the trachea is in the midline. As you all know, compression masses shift the trachea away from the involved side while collapse tends to draw the trachea toward the affected side. These considerations, I feel, exclude pneumonia as a possibility.

D. *Encapsulated empyema* does occur at any age group but the antecedent history of infection is absent. The location is most unlikely and the absence of pleural reaction, I feel, definitely rules this out.

E. *Atelectasis* could occur in this area but here again the absence of tracheal or mediastinal shift is definitely against it. An adenoma or foreign

body would be the most likely etiological agents, and I see nothing to suggest either of these in the x-rays. As you know, atelectasis usually causes a pie-shaped, radiating shadow and not the smooth, rounded mass with which we are dealing today.

F. *Hemangiomas* may be multiple or rarely one massive lesion may be encountered. They are thought to be congenital and seldom cause symptoms.

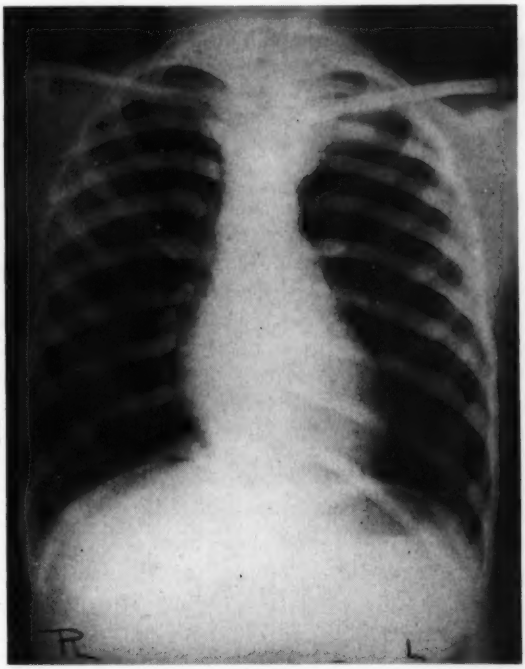


FIG. 1. Initial x-ray showing a smooth rounded mass in the area of left upper mediastinum.

When multiple lesions occur they are located well peripherally and operation is of no benefit. If this shadow represents a massive single hemangioma, a vascular hum undoubtedly would be present. Since the lungs were considered normal to percussion and auscultation, this should be excluded.

G. *Primary lung tumor* is extremely rare in children. Metastases to the lungs occur with lesions located elsewhere, but a primary neoplasm of the lung almost never occurs in this age group. Here again the absence of tracheal shift and the location give added support to excluding this from consideration.

H. *Tuberculosis.* The lungs are clear throughout. Even the hilar areas show little evidence of lymphadenopathy. Tuberculomas, when they occur, usually are smaller. There is no calcium seen along the margins of this lesion and you may remember tuberculomas always occur next to a pleural surface. I feel that tuberculosis is not our problem.

Turning now to lesions occurring in the mediastinum, we first should consider the boundaries and general contents of this area.

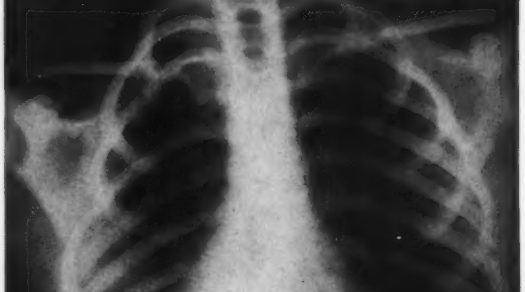


FIG. 2. Post Thoracotomy x-ray showing absence of both the mass and the fourth rib on the left.

The mediastinum is the space located between the medial aspects of both pleura, behind the sternum and in front of the vertebral column. It contains many important structures. It consists of four divisions. The superior mediastinum lies above a line drawn from the junction of the manubrium and the body of the sternum to the body of the fourth intervertebral disc. It contains important blood vessels, part of the thymus, and lymph nodes.

Below this area are the three remaining divisions. The anterior segment lies behind the sternum and in front of the heart. It contains part of the thymus and some lymph nodes.

The middle mediastinum contains the heart and pericardium with the great vessels. The posterior division lies behind the heart and contains the esophagus, trachea, lymph nodes, and the vagus, intercostal, and sympathetic nerves.

II. *Lesions Originating Outside but Extending Into The Mediastinum*

A. *Thyroid or parathyroid* masses are not commonly encountered. They almost always are found in the anterior mediastinum. Adenomas of both organs are rare in this age group. Although they may occur without causing metabolic disturbances, I believe that some of these studies would have been included in the workup had this been seriously considered. There were no masses palpable in the neck or visible on x-ray. The bones which we see on x-ray all appear normal and on this alone, I believe we can rule out a parathyroid adenoma. Because the mass we are explaining is located well posteriorly, I believe these should be ruled out.

B. *Cystic hygroma or lymphangioma* is commonly seen in this area but I have not seen a case with a mediastinal extension which did not have a component in the neck or back. Mediastinal cysts are much less common than tumors and when they occur they rarely cause compression. They tend to conform to the surrounding structures. Mediastinal cystic hygromas usually shift with respirations and with forced inspiration may ride completely out of the chest. These considerations make this an extremely poor possibility.

III. *Lesions Originating in the Mediastinum*

A. *Vascular.* If one saw only the AP x-ray, he could not exclude the shadow as representing a *persistent left superior vena cava.* This is a fairly common finding and produces no symptoms. The same x-ray view is consistent with the condition in which *all of the systemic veins empty into the left auricle.* This is extremely uncommon and in order to sustain life, a coexisting large intra-auricular septal defect must exist. Almost always there are varying degrees of cyanosis accompanying this condition but this was absent here. I feel both of these conditions can be ruled out. A *pericardial cyst* is another possibility which should be considered. I have not seen one presenting in this area. They almost always appear lower and commonly lie close to the diaphragm. They pulsate with the heart and do not cause compression.

B. *Esophageal lesions* are fairly frequently encountered. A *duplicaton* is probably the most common one in this age group. It usually presents on the right for some unknown reason; however, it can present on either side. Varying degrees of obstruction usually accompany this condition. Excision of the duplication usually requires resection of a segment of esophagus. Had this child been subjected to such a procedure, I doubt seriously that he would have been discharged from the hospital two weeks later. *Diver-*

ticula may occur but always cause regurgitation of food which has been ingested hours or days before. *Esophageal tumors* are almost non-existent in this age group and they always produce some degree of dysphagia. I think this group of lesions can be ruled out.

C. *Thymic enlargement or thymoma* always present in the anterior mediastinum. Posterior thymic lesions are out of the question. Many conditions which once were thought to be due to the thymus have been found to represent primary tumors of another type.

D. *Tuberculous lymph nodes* may produce many of the symptoms found in this case such as tracheal compression and erosion. The bleeding would be easily explained by these, but there is no evidence of tuberculosis elsewhere. When these nodes erode into the bronchus there are varying degrees of atelectasis and this is notable by its absence in this case. The location, size, and shape of the mass are definitely against this diagnosis.

E. *Lymphomas* almost always are situated in the anterior mediastinum. *Lymphosarcoma* frequently produces an irregular outline and often other nodes are involved. *Hodgkin's disease* produces blood changes and lymphadenopathy which makes the diagnosis fairly simple.

F. This leaves only two groups of tumors to be considered: the first of these being the *teratomas* and *dermoids* which will be discussed together. They usually are situated in the anterior mediastinum but may occur elsewhere. I see no structure on x-ray suggestive of teeth, bone, or cartilage. If the long strings of mucous previously referred to represent hair, I would say definitely that this lesion is a teratoma or a dermoid. If not, the most logical diagnosis is one of the *neurogenic tumors* which almost always occur in the posterior mediastinum, usually arising from the sympathetic chain, or from one of the intercostal nerves. The various neurogenic tumors cannot be differentiated on x-ray, the two most common ones being the neurofibroma or the ganglioneuroma. They cause compression and are rounded and smooth. The production of a Horner's syndrome is a common accompanying feature. The position of the mass is the usual one for tumors of this type. The dyspnea and cough must have been due to tracheal compression. The bleeding must have resulted from erosion of the mucous membrane. I feel definitely that this is the condition with which we are dealing, and will make my diagnosis one of the posterior mediastinal, neurogenic tumors.

Dr. Robert O'Donnell: Does the smoothness and the spherical shape as seen on x-ray fit in with your diagnosis?

Dr. Marshall Sanford: Yes, it does.

PATHOLOGIC DISCUSSION

Richard J. Waters, M. D.

We are grateful to Dr. Marshall Sanford for his most comprehensive discussion of this case.

The patient was seen in consultation by representatives of the radiology and thoracic surgery departments. Flouroscopic examinations revealed the mass to be posterior and smooth in outline. It appeared fixed and did not pulsate. It did not impinge upon the trachea or the esophagus and there was no erosion of the rib cage or vertebrae. Following lengthy consideration, it was decided that the most likely diagnostic possibility was a tumor mass of the left posterior mediastinum.

Following bed rest and transfusions of whole blood, a thoracotomy was performed. Access to the area was gained by the subperiostial resection of the fourth rib on the left. Following incision of the pleura, the contents of the mediastinum were viewed from the left and a bilobed, firm, smooth, tumor mass measuring 5.5 cm. by 3.5 cm. was seen to occupy the sites of the third and fourth ganglia of the thoracic sympathetic chain. There was distinct tissue continuity between this mass and the sympathetic ganglia immediately above and below it. There was no connection with the tracheo-bronchial tree. The tumor was enucleated and the surgical procedure was completed without complication.

Further examination of the gross specimen revealed the cut surface to be smooth and irregularly red and yellow. There were intermingled grey fibrous tissue trabeculae and several hard granules of calcium like material. The general architecture suggested nervous tissue. Histologically, the characteristic morphology was the occurence of great numbers of "ganglion" type cells which varied greatly in their stage of development and were embedded in a fibrous stoma of Schwann cells. The process was believed to have arisen in a sympathetic ganglion and the histologic diagnosis was ganglioneuroma.

Ganglioneuromas of peripheral nerve cells and their processes occur rather frequently. These tumors are slowly growing and cause symptoms in direct proportion to their size or proximity to important structures. Ganglioneuromas of peripheral nerves appear anywhere in the body but are more commonly found along the paravertebral lines in the thorax or abdomen. Malignant change is uncommon, but it does occur.

I am happy to report that four years and eight months after operation this child is in good health and doing well in school. There are no signs or symptoms of metastasis.

McFarland, J., and Sappington, S. W.: Am. J. Path. **11:** 429, 1935.